



IMAGE

Aspects of a neglected tropical cardiomyopathy: Endomyocardial fibrosis

Fibrose endomyocardique tropicale : une cardiomyopathie encore mystérieuse

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We present the case of an 11-year-old boy who came from the north of Maputo in Mozambique, an area endemic for endomyocardial fibrosis. He presented with giant ascites secondary to a fibrotic endocardial biventricular thickening that was far more prominent on the right side. Chest X-ray showed severe cardiomegaly secondary to a giant right atrium (RA). Echocardiography confirmed that the fibrous right ventricle (RV) was totally reduced at its infundibulum with an aneurismal right atrium (apical four-chamber view) (Fig. 1). Unfortunately, treatment was medical only, without the possibility of cavopulmonary derivation because of the left fibrous extension with severe diastolic dysfunction of the left ventricle (LV).

Comment

Endomyocardial fibrosis is an endemic disease in certain tropical areas. Nearly 70 years ago, Arthur Williams noted unusually large patches of fibrosis at autopsy in two Ugandan patients with severe mitral incompetence. At this time, Williams was perplexed and ascribed the disorder to syphilitic myocarditis. Eight years later, in 1946, Bedford and Konstam reported at a meeting of the British Cardiac Society: ‘‘obvious and extensive subendocardial fibrosis, with fibrous areas resembling shallow infarcts in the ventricles, adherent to which were organized antemortem clots’’. The significance of these lesions was recognized by Davies in Uganda, who in 1947 described in detail the pathological features of the disease and designated the dense fibrous plaque of the mural endocardium as endomyocardial fibrosis.

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Figure 1. Clinical, radiographic and echographic aspects of a neglected tropical cardiomyopathy: endomyocardial fibrosis.

Endomyocardial fibrosis remains endemic among children who belong to poor sections of society in certain tropical areas. Rare clinical epidemiological studies have shown a heterogeneous distribution of the disease, with environmental, local or inherited (ethnicity) factors favoring its development. The pathogenesis of this condition remains unknown. Several hypotheses have been proposed and explored, including cardiotoxicity of the eosinophil, infectious agents, autoimmune processes, genetic predisposition, ethnicity, diet and geochemical factors. Although eosinophilia is seen in most cases of endomyocardial fibrosis during the initial inflammatory process and histological endocardial lesions are similar to the hypereosinophilic syndrome observed in industrialized countries, eosinophil cell toxicity is not accepted by all researchers. In any case, according to this theory, the origin of hypereosinophilia remains obscure. Ascites with little or no peripheral edema is the most demonstrative clinical feature of the disease, regardless of which ventricle is affected. It is still unclear whether

these clinical aspects of endomyocardial fibrosis are linked only to strong right cardiac pressure or are proof of a systemic disease. Recently, has been published in the *New England Journal of Medicine*, the first population study of endomyocardial fibrosis in a rural area of Mozambique using a systematic transthoracic echocardiography [1]. By using echocardiography, Ana Olga Mocumbi et al. were able to detect early, asymptomatic stages of the disease, and estimated the overall prevalence up to 19.8% in this endemic area (Inharrime district in the north of Maputo). The authors concluded that "these findings may aid in the study of the pathogenesis of the disease and in the development of new management strategies".

Reference

- [1] Mocumbi AO, Ferreira MB, Sidi D, Yacoub MH. A population study of endomyocardial fibrosis in a rural area of Mozambique. *N Engl J Med* 2008;359(1):43–9.